CLINICAL PROCEEDINGS

OF THE CHILDRENS HOSPITAL

13th and W Streets, Washington 9, D. C.

Vol. VI *May 1950* No. 6

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Published monthly by the Staff. Cases are selected from the weekly conferences held each Sunday morning at 11:00 A.M., from the Clinico-pathological conferences held every other Tuesday afternoon at 1:00 P.M., and from the monthly Staff meetings.

This bulletin is printed for the benefit of the present and former members of the Attending and Resident Staffs, and the clinical clerks of Georgetown and George Washington Universities.

Subscription rate is $1.00 per year. Those interested make checks payable to "Clinical Proceedings Dept.," The Children's Hospital, Washington, D. C. Please notify on change of address.

Entered as second class matter November 21, 1946 at the post office at Washington, D.C., under the Act of March 3, 1879. Acceptance for mailing at special rate of postage provided for in Section 538, Act of February 28, 1925, authorized January 17, 1947.

THE PROBLEM OF TONSILLO-ADENOIDECTOMY IN CHILDHOOD

Allan A. Walker, M.D.*

Tonsillo-adenoidectomy in children presents a problem primarily because the function of these lymphoid structures is not well defined nor understood.

A brief review of the anatomy, physiology, and pathology of these structures is presented in an attempt to assign some function to them. The tonsils include the tubal tonsils, the ring of tissue surrounding the eustachian tube, the nasopharyngeal tonsils (adenoids), the faucial or palatine tonsils and the laryngeal tonsil. This ring of lymphoid tissue is known as Waldeyers Ring. Embryologically, these structures develop and atrophy in a cephalo-caudal direction. The enlargement of the tubal tonsil and nasopharyngeal tonsil begins after birth and reaches its peak after the second and third years. At puberty, the involution of these structures becomes accelerated and by eighteen to twenty years of age the adenoid has normally disappeared and the tubal tonsil has become smaller. The accelerated development of the faucial or palatine tonsil begins later in infancy than the adenoid, and usually completes its involution by the fourth decade of life. The lingual tonsil which comprises the fourth link in Waldeyers Ring exhibits its maximum development about this time, that is, in the fourth decade.

The epithelium comes into direct contact with the lymphoid tissue comprising Waldeyers Ring, a fact which distinguishes it from the other lymphoid tissue of the body. The tonsillar tissue is composed of a parenchyma and secondary nodules. These secondary nodules are not present at birth but are rapidly developed afterward. Guinea pigs raised under sterile conditions do not develop these nodules at two months, a time when they are normally present in controlled specimens. Daily bacterial injections into newborn rabbits caused premature development of these nodules at three weeks. There is evidence that these nodules play a part in the process of immunization and are important in the production of antibodies. Clinical observation has substantiated this fact. Reports in the literature prove that children who have had early removal of the faucial tonsil are more susceptible to respiratory infection.

The efferent drainage of Waldeyers Ring into the regional cervical lymph nodes is a well known anatomical feature.

Chronically diseased tonsils may act as nidus of infection. This infection spreads out to involve the surrounding tissues. Sometimes the infection

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may be so immediately generalized throughout the lymphoid structures that the tonsillitis must represent only a local manifestation of systemic disease. Tonsillitis, per se, is not an infection of the surface of the tonsil, but an infection which extends throughout the entire substance of the tonsillar tissue.

In summary, accumulated evidence indicates that the tonsils assist in the development of immunity in childhood, but probably have no vital function. Secondary involvement of adjacent vital tissue is more important than the primary disease of the structures of Waldeyers Ring. In children the eustachian tube is shorter, more horizontally disposed, and relatively lower in its position in the nasopharynx, and so more vulnerable to infection. Surrounding the eustachian orifice is the tubal tonsil. Adjacent to the tubal tonsil is the mass of the adenoid, often hypertrophied to such a degree that it obstructs the opening of the tube. This anatomical disposition may cause recurrent attacks of acute suppurative otitis media, chronic otitis media, or chronic obstruction of the eustachian tube leading to partial deafness.

The intimate relationship of the adenoid to the posterior nares vitally effects the nose and its related structures. Infection of the adenoid may pass by direct extension to the mucous membrane of the nose and thence into the sinuses. By causing mechanical obstruction to the posterior nares, the adenoid brings about stasis of the secretion of the mucous membrane and institutes abnormal conditions in the nose leading to infection. The picture of a child with an obstinate adenoid condition is a familiar one. Externally the nose is underdeveloped, the upper teeth protrude, the palate is arched, the nasal turbinates are boggy, and excessive mucoid secretion is present in the nose. The child is underweight, nervous, and restless at night. The failure of the mucosa of the nose to moisten and warm the inspired air causes irritation to the tracheo-bronchial tree. Upon the already irritated lower respiratory mucous membrane is superimposed the irritation of a continuous postnasal discharge, bathing this mucosa in pus.

Removal of diseased tonsils depends upon thorough examination of the nose and throat. The size of the faucial tonsils and the presence or absence of inflammation is taken into consideration. The adenoid mass and tubal tonsils must either be palpated or visualized with a naso-pharyngeal mirror or pharyngoscope. The conditions of the related structures e.g., the nose, sinus, ears, chest, and cervical nodes must be observed. Frequently the otolaryngoscopist must be guided by the recommendation of the pediatrician who has had the opportunity of seeing the lymphoid tissue during repeated episodes of acute infection. He has observed the effects of these episodes on the related structures and on the general health of the child.

There are three aims of therapy in diseased tonsillar structures:

1. Elimination of infection.
2. Correction of the secondary effects of the disease upon adjacent tissue.
3. Reestablishment of the well being of the child.

There is no age barrier to surgical removal of the adenoid mass. The effects of complete mechanical obstruction on a young child are so profound and serious, the operation so minor, the post-operative course so mild, and the results so satisfactory that adenoidectomy may be performed at any time. If the adenoid mass is causing only partial obstruction, it is often possible to bring about sufficient decrease in its size simply by eliminating nasal infection. Correction of a nasal allergy often results in a spectacular decrease in the size of the adenoid mass. The tubal tonsil should not be removed surgically. Again attention should be paid to the elimination of infection and allergy in order to bring about decrease in its size and re-establishment of patency of the tubes.

Recently radium and x-ray therapy has been adopted and used; however, these measures may be utilized with too much enthusiasm and indiscrimination. The rays destroy lymphocytes undergoing mitosis. The administration of divided doses materially reduces the number of lymphocytes in the infected tissues. The acceptable method of therapy consists of the application of 50 milligrams of radium placed in the nasopharynx for twelve minutes on each side at intervals of three weeks, for a total of three applications. In the past several years it has been shown that this dose may be sufficient to ultimately stimulate the growth of neoplasms in this area. Certainly it is unwise to apply radiation therapy until all else has been tried.

Tonsillo-adenoidectomy is best performed under the conditions of minimal infections. It is sometimes impossible to achieve a period of quiesence and it has been found to be relatively satisfactory to operate on the child who is well saturated with sulfadiazine and penicillin. On two occasions I have seen children with an acute upper respiratory obstruction from acute tonsillitis and have had to make the choice between tonsillectomy and tracheotomy. With the use of these drugs no ill effects were produced from electing to perform the tonsillectomy rather than the tracheotomy.

In preparing the child for surgery the psychic element is most important. It is at best a disagreeable episode. The events leading up to actual operation should be discussed with the child. It is most reassuring to the child to have the mother stay and it is my preference that the parents be with the child during induction of anesthesia.

In conclusion, I would like to emphasize that a tonsillectomy is a major procedure and that it produces a profound physiological upset in the child. Tonsillectomy should not be undertaken lightly and if possible should be avoided during the early years of life.

Edgar Copeland, M.D.*

I have been requested to discuss the indications and contraindications for tonsillectomy and adenoidectomy from the standpoint of the pediatrician. Such a discussion immediately precipitates many controversial questions. I am obliged, in consequence, to employ the first person, for I may not find universal acceptance of my own ideas. I think I may say however, that my thoughts on the subject reflect the considered opinions of most pediatricians, especially those who do not do their own surgery, and at least one oto-laryngologist, by name Allan A. Walker, M.D.

The pediatrician is constantly being consulted to pass upon the merits of such cases among the children who have been under his own supervision over a period of time, as well as children whom he is seeing for the first time. I think all will agree as to the greater value of his judgment in the former group, while in the latter there might be some difference of opinion, especially if the surgeon has had pediatric training.

As an outline for any discussion I will say that the general indications for tonsillectomy and adenoidectomy may be divided into two groups. In the first group are the cases involving mechanical obstruction, to the drainage and ventilation of the accessory nasal sinuses, the eustachian tubes and to respiration and deglutition. In this group will be the occasional instance requiring emergency attention, as in the case mentioned by Dr. Walker, where a tonsillectomy was substituted for a tracheotomy.

In the second group, by far the larger one, will fall the much more common indications for operation. These indications are as follows:

1. Frequent, recurring attacks of tonsillitis.
2. Chronically infected tonsils, with or without septemic involvement.
3. Tonsillar abscess, especially when recurring.
4. Chronic sinusitis.
5. Cervical lymphadenitis with impending suppuration.
6. The nephritides.

In arriving at a decision in a particular case we are forced to appraise the importance of the functions of the lymphoid structures in the naso-pharynx and weigh advantages removal of against the loss of such functions in the diffuse mechanisms of the body.

This lymphoid assembly, known as Waldeyers Ring, the anatomy of which has been seen well visualized for us by Dr. Walker, is an important part of the mechanism of defense against infection. This function is not to be too lightly regarded, especially in the early years before the development of immunities that take over much of this responsibility later on. The accelerated growth of lymphoid tissue throughout the body up to the age of

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twelve years, so well illustrated in Scammon's graphs, strongly suggests a vital role in human physiology, even though it is not well understood. It is significant that an hyperplasia of adenoid nodes is often precipitated by their removal in the very young patient, resulting in a reversal of the dramatic results that have followed the initial surgery. Also it may be said, that the incidence of bronchitis and lower respiratory infections replacing tonsillitis, after tonsillectomy and adenoidectomy is sufficiently common to suggest that the operation has removed some sort of a barrier to such infection.

In spite of what I have said about the diffusive function of lymphoid tissue, there will arise from time to time, very definite indications for its sacrifice. There is undoubtedly a genetic factor in the size of tonsils, some individuals being more liberally endowed with such tissue than others. Environmental factors also have a bearing. An abnormal amount of lymphoid tissue however, suggests the hyperplasia resulting from the stimulation of recurring inflammatory bouts. It is often difficult to determine that the tonsil is infected, but it is reasonable to assume that such is the case when there is a failure of the structures to regress to something approaching their original condition after recovery. Associated cervical lymphadenitis increases the probability of chronic infection.

Before the era of general immunization diphtheritic infection of the tonsils often presented a tedious problem. I have not seen the tuberculous tonsil, probably because of a lack of awareness, but in some statistics it has not been an uncommon infection. Since it is observed in the young without demonstrable evidence of the disease elsewhere and since the prognosis is said to be good, operation would seem to be imperative.

To have observed the favorable results of tonsillectomy and adenoidectomy in the child, with recurring colds, obscure fever, the abdominal pains so characteristic of tonsil infection, anorexia, unsatisfactory symptoms, is to be all but convinced of the value of operation. It should not be overlooked however, that much, if not all, of the favorable response may be the result of coincidental factors in a new regimen.

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REMOVAL OF FOREIGN BODIES FROM THE STOMACH AND UPPER INTESTINAL TRACT BY MEANS OF A SMALL MAGNET

Case Report No. 182

Philip A. Caulfield, M.D.

E. G., a two year old white female sent into Children's Hospital by Dr. Robert Hightower with a history of having swallowed a bobbypin shortly before admission. X-ray examination revealed the bobbypin to be lying in the stomach in a transverse position. The parents were quite concerned about the child's condition and were very anxious that an attempt be made to remove the pin. They were informed that the pin would probably pass through the intestinal tract without injury to the child, but that it would take some time before it would pass through the entire intestinal tract. It was suggested that a small magnet could be attached to the end of a tube, and since the object was steel, would probably adhere to it. They readily agreed to this suggestion. A small magnet was then attached by means of a piece of heavy twine, to the end of a Levine tube. The child was gagged and the magnet along with the tube was inserted into the stomach. The child was immediately placed under a Fluoroscope which revealed that the bobbypin had already adhered to the magnet. The tube along with the magnet and attached bobbypin was then withdrawn. The entire procedure took less than ten minutes.

DISCUSSION

Small foreign bodies containing steel or iron, lodged in the stomach or upper intestinal tract, can easily and safely be removed by means of a small magnet attached to the end of a Levine or Miller-Abbott tube. This magnet, measuring about one-half inch long and a quarter of an inch wide, rectangular in shape, can easily be obtained from any novelty shop or five and ten cent store. Tied securely to the end of the tube by means of a piece of thread, it can be inserted into the stomach without any great discomfort to the patient. This holds true for small children as well as for adults. The magnet is of sufficient strength to remove any object located in the stomach, almost as soon as it enters the stomach. On many occasions, the object in the stomach will be attached to the magnet before a Fluoroscope can be placed over the patient to watch the progress of the removal. The magnet is also sufficiently strong so that if the object is longer than the magnet and lies transversely as it strikes the esophagus, the magnetic field will allow it to be turned longitudinally, and easily slide through the esophagus. Most

straight pins, bobbypins, tacks, small nails and other objects contain steel, and can readily be removed. However, most safety pins are made of brass and can not be removed. No coins, except the zinc penny manufactured during the war, can be removed because they do not contain steel. However, the zinc penny contains a large percentage of steel and can readily be removed.

Most foreign bodies that lodge in the stomach will pass into the intestinal tract and be extruded without causing the patient any particular harm. However, there is a great deal of anxiety on the part of the patient, his parents and the physician, as to whether or not the object is going to pass. Frequently great expense is entailed by serial x-rays to watch the progress of the foreign object through the intestinal tract. The use of a magnet for removal of these objects, shortly after they are swallowed, alleviates all these fears, as well as the necessity for an Abdominal Exploration for those objects that will not pass, and thus reduce expenses for taking care of this type of patient.

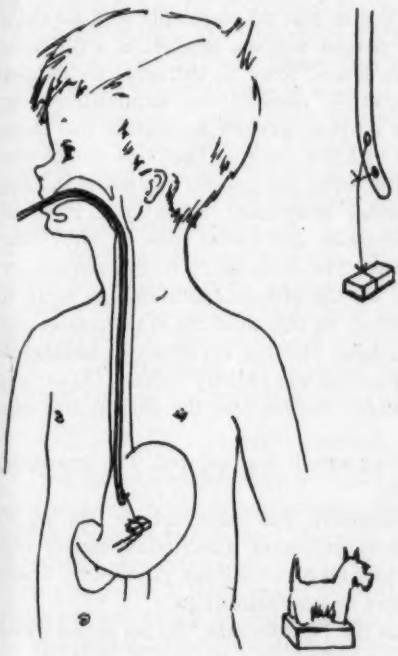

FIG. 1. The magnet has been attached to the levine tube by a piece of dermal thread and inserted into the child's stomach. A bobby pin has attached itself to the magnet.

The removal of foreign bodies from the intestinal tract by means of a magnet, is not a new procedure. It has been advocated from time to time in the literature, and has been very successfully tried on numerous occasions. At a recent meeting of the American Pediatric Society in Baltimore, Maryland, there was a large exhibit that showed numerous objects having been removed by the use of a magnet from the intestinal tract.

Occasionally, foreign bodies entering the stomach and intestinal tract do become lodged and penetrate the wall of the gut. By using a magnet, the frequency of this occurrence will be reduced, and these patients will not be subjected to the hazards of peritonitis and an abdominal operation.

DIFFUSE HEMANGIOMA OF THE LIVER

Case Report No. 183

Joseph M. Lo Presti, M.D.
Paul Kaufman, M.D.
Francis J. Troendle, M.D.

B. W. 49–10138

B. W.,* a two and one-half year old white male, was hospitalized on August 15, 1949 with a chief complaint of semi-consciousness of two days' duration. He had been apparently well until five days before admission at which time he began having intermittent episodes of vomiting. For two days prior to hospitalization the vomiting had ceased; however, there was marked anorexia. Throughout the entire illness extreme irritability and insomnia had been present. The bowel habits had never been normal; a mild diarrhea had persisted almost from birth. At the onset of the present illness this usual picture changed to one of constipation.

His mother was grava ii, para ii and had an uncomplicated prenatal course during this pregnancy. The patient was the product of a full-term gestation; labor had lasted for five hours. Prior to delivery the mother had been sedated with rectal pentothal; demerol was administered for analgesia. At the time of delivery ethylene-oxygen anesthesia was instituted, and the infant was delivered with low forceps. The birth weight was seven pounds one ounce. Shortly after birth, the patient was noted to have irregular respirations with inspiratory intercostal retractions. Physical examination revealed impaired resonance and coarse rales at both lung bases. The patient was a poor feeder. The irregularity in the respiratory rate necessitated the use of oxygen. On the fifth postnatal day an x-ray of the chest was reported as being normal. At this time the Wassermann was negative; the hemoglobin was 11 grams; and the erythrocytes numbered 3,490,000 per cubic millimeter. A urinalysis was entirely normal. On oxygen therapy alone there was gradual improvement, and the patient was discharged on the seventh day of life.

The mother, father and older sibling were living and well. The remaining family history was non-contributory.

Growth and development were retarded. The child had just started to walk and speak a few months prior to admission. The feeding history was adequate. Routine immunizations had been carried out in infancy. There had been no previous serious illnesses or hospitalizations.

On admission the temperature was 102 F; pulse rate, 120 per minute; and

* The patient was admitted to the private service of Preston A. McLendon, M.D.

the respiratory rate, 46 per minute. The patient was a well developed white male with marked dyspnea who appeared to be in extremis. He whined constantly but did not respond to nociceptive or proprioceptive stimuli. The skin was dry, pale, and cold. No petechiae or rashes were present. The eyes were enophthalmic and glazed. The pupils were dilated but the funduscopic examination revealed no abnormality. The conjunctivae and all mucous membranes were very pale. There was no nuchal rigidity. The lungs and heart were normal. The abdomen was distended and covered with a visible venous pattern. Borborygmus was absent. The liver was palpable approximately 10 cm. below the right costal margin. It was firm in consistency, and the edge was smooth and rounded. A mass which was thought to be the spleen was palpated about 10 cms. below the left costal margin and possessed the same consistency as the liver. The remaining physical examination including the neurological was normal.

Admission urinalysis revealed 300 mgm. per cent of albumin, a three plusacetonuria, and the sediment contained innumerable red blood cells. The hemoglobin was 7.5 grams and the erythrocytes numbered 2,500,000 per cubic millimeter. The leukocytes were 10,000 per cubic millimeter with a normal differential count. There were 185,000 platelets per cubic millimeter. Bone marrow aspiration was reported as normal.

On admission a phlebotomy was performed and continuous intravenous fluids were administered. During the next twenty-four hour period the patient received 750 milliliters of 5% dextrose in normal saline, 550 milliliters of whole blood, 500 milliliters of plasma and 150 milliliters of normal saline. Wangensteen drainage was instituted and a "coffee ground" material was aspirated from the stomach. On the day after admission blood was noted to be oozing from the site of the phlebotomy. The "pin-prick" from which a blood sample for hemogram had been taken continued to bleed for an hour despite the use of local pressure. The temperature rose steadily to 106 F. and the pulse rate dropped precipitously to 82 per minute. The patient expired approximately twenty-five hours after admission.

PATHOLOGICAL DISCUSSION

Post mortem examination revealed a well nourished and developed two and one-half year old white male whose length was 98 centimeters and weight was 13,440 grams. The skin was normal, no rashes, bruises, or hemangiomata being present. The abdomen was slightly distended and the liver was palpable three fingers below the right costal margin. A few dilated venules were visible over the abdomen.

The peritoneal cavity contained about 75 milliliters of clear, yellow fluid and there were no adhesions of the serous membranes.

The only organs showing pathologic changes were the lungs, liver, kid-

neys, intestine, and brain. The lungs grossly were slightly heavier than normal although they appeared smaller. On cut section, portions of the lung parenchyma appeared more mottled, firmer, and less crepitant than the remainder, suggesting pneumonia and atelectasis.

The liver was very large, extending 7 centimeters below the midline and the right costal margin. Its lower edge was rounded and its color was a mottled red, green, and black. The surface was smooth and tense to palpation. Aspiration of the organ revealed free flowing blood. When sectioned, the liver resembled a sponge which had been soaked in blood. The hepatic parenchyma was visible as a network of tissue forming venous lakes of various sizes. No large areas of hepatic destruction were present, however.

There were several small ecchymotic areas in the mesentery of the small bowel along with many reddened lymph nodes. A hematoma measuring 2 x 3 centimeters was present in the serosa of the sigmoid but did not communicate with the mucosa or the lumen.

The right kidney was slightly larger than the left and on cut section, a hemorrhagic area was visible in the region of the renal calyces. The cortex and medulla were normal. The right ureter contained a partially organized clot in its lumen about 4 cm. from the uretero-vesicle junction.

The surface of the right parietal area of the brain was covered with a purple clot of blood measuring 4 by 5 centimeters which was easily removed by wiping. The brain on coronal section showed only slight congestion. The cisternal fluid was grossly bloody.

Histologically, the lung revealed alveolar wall thickening with an infiltration of polymorphonuclear cells in the walls and alveolar lumena. An exudate of fibrinous and fatty material was present in the lumena of many alveoli, indicating aspiration.

Microscopic section of the liver revealed a large number of blood filled spaces, varying in size from capillaries to large sinusoids. Many of these spaces were lined by endothelium varying from cuboidal type in the small vessels to pavement type in the large sinusoids. Many large groups of capillaries were present. There was no evidence of an abnormal hyperplasia of the endothelium lining the blood filled spaces.

Precipitated fibrin and thin strands of fibrous tissue lined many of the blood filled spaces. The erythrocytes in many areas had degenerated, the resulting debris having assumed an amorphous orange colored appearance. The precipitated material had formed columns in several portions of the section, separated by fibrous tissue strands. Round cells and polymorphonuclear leukocytes had infiltrated into portions of the parenchymal tissue which was undergoing necrosis.

The hepatic cells showed the results of compression by the extravasated blood and the marked vascular engorgement. In some areas, relatively

normal hepatic cells were visible but in other sections only a faint outline of the liver cells was visible, each enclosing a pyknotic nucleus. The majority of the polygonal cells had undergone vacuolization and increased granularity, although the nuclei appeared normal.

In the regions of the hepatic trinities, a marked compression and distortion of the portal venules and bile ducts had occurred.

The kidneys, histologically, revealed marked hemorrhage beneath the mucosa of the renal pelvis but the parenchyma of the kidney was uninvolved except for an occasional collection of lymphocytes and tubular epithelial degeneration.

The pathologic diagnoses were:

1. Massive hepatic hemangioma (cavernous type)
2. Hemorrhage
   a. subarachnoid space
   b. intestinal wall
   c. renal pelvis
3. Atelectasis, bronchopneumonia, secondary to aspiration of foreign material.

Cause of death: Subarachnoid hemorrhage due to clotting defect secondary to hepatic insufficiency.

DISCUSSION

Tumors of the liver are extremely uncommon in infancy and childhood, but when in the rare instances one is encountered, it is more apt to be in the form of the primary, malignant hepatic tumor. Thus, in 1946, Videbaek[1] reviewed the literature and collected a series of thirty cases of hemangio-endotheliomas (hemangio-endothelio-sarcomas) of the liver and added one of his own. Of interest in Videbaek's collection of cases was the age incidence of this condition. Fourteen patients were in the 0–5 year age group while the remainder occurred between the ages of twenty and eighty years. The fastigium in this older age group was the fifth decade of life. Apart from the somewhat more rapid course of the disease (a peculiarity of neoplastic disease in childhood), there seemed to be little difference between the clinical pictures of both groups. There was no histological difference in the construction of the tumor, but, unlike children, adult patients frequently exhibit cirrhosis of the liver.

Herxheimer[2] in 1930 was able to collect only 439 cases of primary carcinoma of the liver and sixty-six cases of primary hepatic sarcoma in his review of the literature up to that time. Ten per cent (forty-four) of the cases had occurred among children below ten years of age, and 5.1 per cent (twenty-seven patients) were between the ages of ten and twenty. In the period between 1917–1929, Pack and Le Fevre[3] found only twenty-eight

patients suffering from primary carcinoma of the liver out of the 16,565 patients who had passed through The Memorial Hospital during that time. The youngest patient in this series was twenty-two years old. In a review of the medical literature on primary hepatic carcinoma in childhood, Steiner,[4] in 1938 could accept only seventy-five of the one hundred and five published cases as being correctly diagnosed. Fifty-three per cent (forty) of these children had acquired the disease before two years of age.

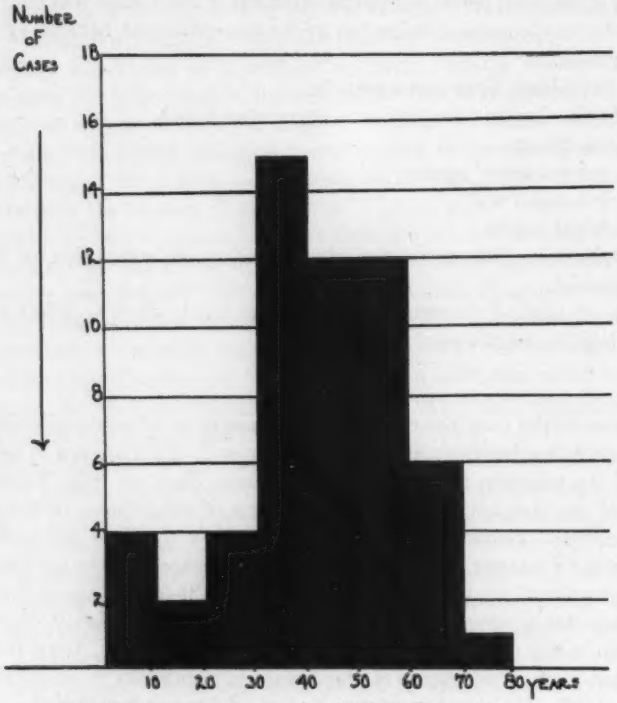

TABLE I AGE DISTRIBUTION OF 58 PATIENTS WITH HEMANGIOMA OF LIVER.

Although primary sarcoma of the liver is more uncommon than primary hepatic carcinoma, twenty-three per cent of the sixty-six cases of sarcoma discussed in Herxheimer's publication[2] occurred in the 0–10 year age group as compared to ten per cent of the carcinomas. This appears to be in accordance with the general observation that sarcomas occur with comparative frequency in childhood.

Of the benign tumors, hepatic hemangioma, along with adenomas and

cystic tumors, occur with the most frequency. Fiddler and Madding[5] in 1949, reported a case of hemangioma which brought the total number of recorded cases to seventy-two. The patient reported was a sixty year old white female.

FIG. 1. Enlarged, smooth liver surface with rounded edge extending 7 centimeters below the right costal margin and midsternal line.

Cystic disease of the liver, a common benign hepatic tumor, is occasionally associated with cystic kidneys.[6] Non-parasitic cysts are rarely encountered and may reach considerable size; in some instances the cyst is extra hepatic, being attached to the liver by a fibrous pedicle.[7]

Metastatic neoplasms of the liver in childhood originate chiefly from neuroblastomas. In young infants, especially, they may produce marked hepatomegaly. Neuroblastoma may occur and metastasize in uterine life. In 1930, Hagstrom[8] recorded the occurrence of fetal dystocia due to metastatic neuroblastoma of the liver.

Because hepatic new-growths are quite rare in the young patient, a good deal of confusion exists. In addition, the multiple terminology and the differences in the histo-pathological interpretations of necropsied and operative material adds to the confused state of thinking. With an intent at clarification a classification of the more commonly occurring new-growths

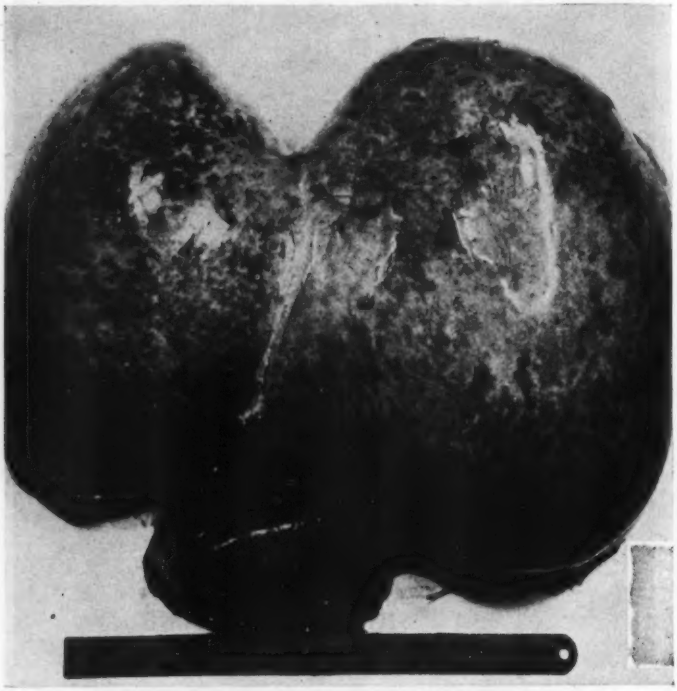

FIG. 2. Liver and spleen. The smooth, rounded, glistening surface of the liver showing mottled areas of red and black discoloration. This organ weighed 1191 grams, twice normal weight.

in children has been attempted:

I. Primary
  A. Benign
    1. Adenomas
      a. Derived from liver cells.
      b. Derived from epithelium of bile ducts.
    2. Cystic tumors
      a. Cystic dilatation of adenomas derived from epithelium of bile ducts.

b. Congenital solitary non-parasitic cysts.
c. Congenital multiple cysts.
3. Hemangiomas
4. Fibromas
5. Hamartomas
6. Teratomas

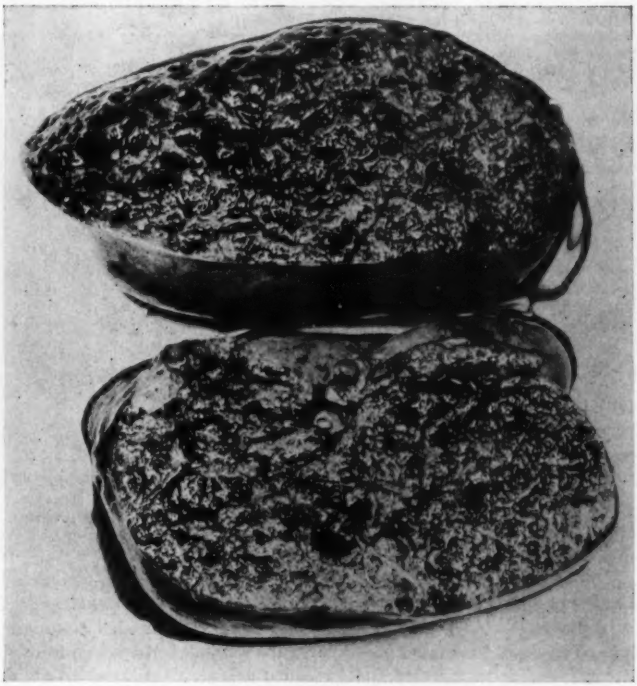

FIG. 3. Cut section of the liver showing the sponge-like appearance of hepatic tissue forming large and small venous lakes.

B. Malignant
1. Carcinomas
2. Sarcomas (including endotheliomas)

II. Secondary or metastatic
A. Neuroblastoma
B. Embryoma of Kidney (Wilm's)

In a review of the medical literature the authors were able to locate seventy-five reported cases of hemangioma of the liver. The ages were noted in fifty-eight of these patients (Table 1). This disease entity is certainly

more common in adults, particularly between the third and seventh decades of life, and has its highest occurrence between thirty and sixty years. Only four cases have been reported in childhood and two of these were in infants. Hendrick,[9] in 1948, reported a case in a newborn. It is of interest that in both of these instances there was a fatal termination caused by a rupture of the hemangioma with resultant massive intra-abdominal hemorrhage. In sixty-one of the cases reported, the sex was noted. There were fifty-one

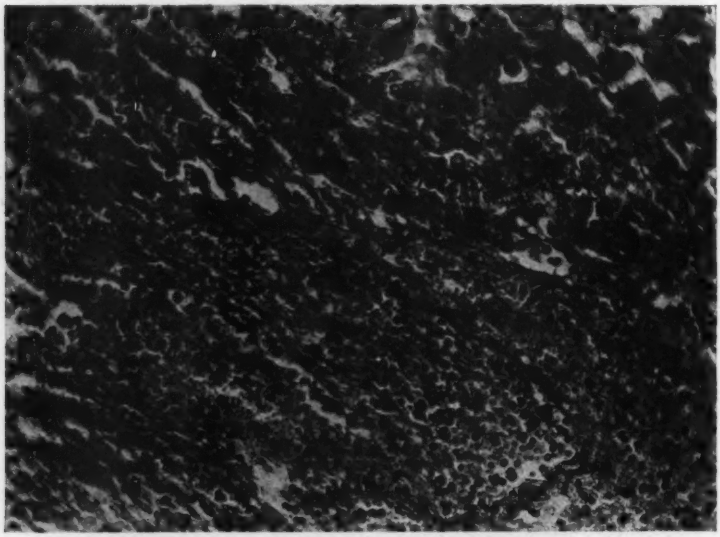

FIG. 4. Photomicrograph revealing recognizable hepatic tissue in the upper right portion of the picture. Large lakes filled with blood and lined by normal endothelium cells are visible in the lower portion of the slide. This portion also shows numerous leukocytes which have infiltrated the area. In the extreme left lower part of the picture one can visualize compressed and distended polygonal cells, which are scarcely recognizable.

males and ten females. There has been no adequate explanation given for the predilection of this condition for males and its more common appearance in adults. Three factors may play a part:

1. The tumor is a slow growing one and does not produce symptoms for many years.

2. Hormonal stimulation may cause a rapid proliferation in adulthood. This theory is given more credence when one realizes that many benign skin hemangiomata become malignant after adolescence.

3. The occurrence of congenital hemangiomata in infants may be explained by placental transmission of these activating hormones.

Hemangioma of the liver may be classified into three different types on the basis of gross pathology:

1. Single or cavernous hemangioma:[11, 12, 13] by far this is the most common type.

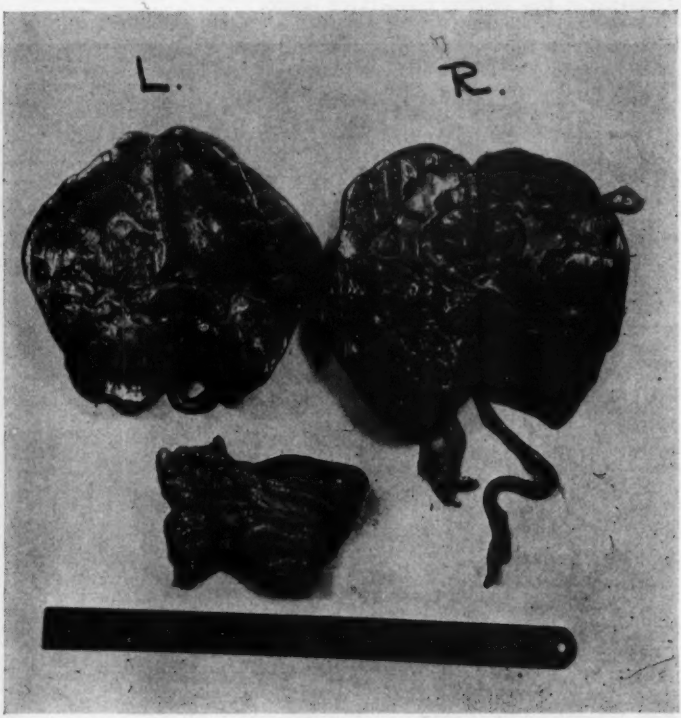

FIG. 5. Cut section of the right kidney revealing hemorrhage in the renal calyces. A small clot is visible at the distal end of the ureter. The left kidney appears normal. Lying below the kidney is a section of the sigmoid colon revealing normal mucosa and a hematoma in the mesentery.

2. Diffuse cavernoma:[14] in which there are numerous varied-sized hemangiomata disseminated throughout the liver.

3. Pedunculated hemangioma: this type may reach tremendous size. Peck,[15] in 1929, reported the surgical removal of a pedunculated hepatic hemangioma which weighed three pounds fourteen ounces.

Histologically the walls of the hemangioma consist of elastic and smooth muscle fibres with an endothelial lining. The gross connective tissue bands may contain atrophic liver cells, bile ducts, or hemosiderin. Thromboses may be encountered in some of the venous sinuses of the tumor.

Various hypotheses concerning the etiopathogenesis of hemangioma of the liver have been advanced:

1. Hanot's theory of dilation of the capillaries of the acinus.
2. Derivation from dilated portal vessels following a primary inflammatory disease of the hepatic connective tissue.

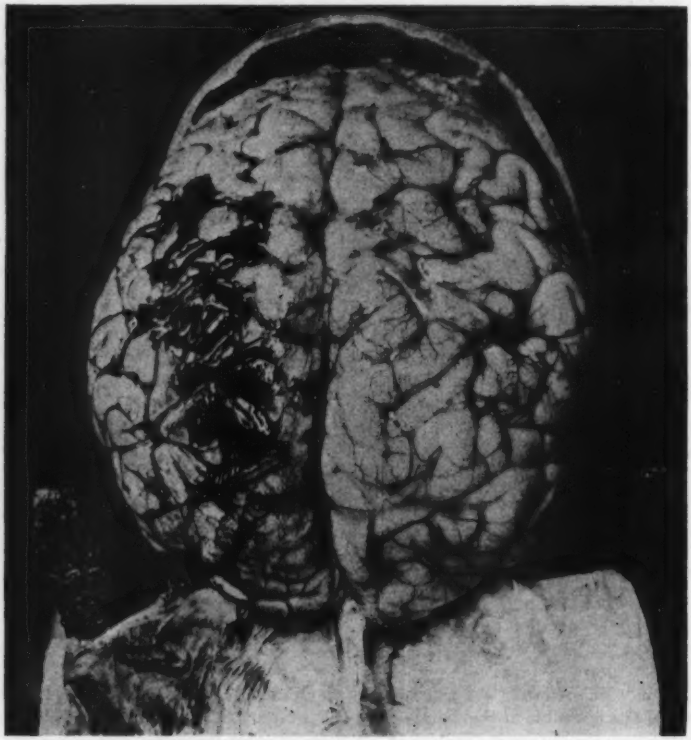

FIG. 6. Surface of the brain after removal of the calverium and meninges revealing a hemorrhagic area covering a portion of the left parietal lobe.

3. True neoformation resulting from developmental disturbances.
4. Derivation from the portal and suprahepatic venous systems.
5. Primary malformation of hepatic tissue, consisting of capillary telangiectases which are transformed into cavernomata by thickening of the walls.
6. Capillary dilatation and hepatic cell compression following biliary obstruction.

The diagnosis of hemangioma of the liver is very rarely made preoperatively or during life. In the seventy-five recorded cases the correct diagnosis was made in four patients. A tumor of the liver was suspected in nine additional cases. Pathognomonically there is a blowing murmur over the tumor, and volume changes of a spontaneous character or after manual compression. In the remaining patients hydatid cyst of the liver, ovarian cysts, pancreatic cysts, malignancy of the gastrointestinal tract, hydrops of the gall bladder, a mobile kidney, cholecystitis, appendicitis, mesenteric cyst, perforation of a viscus, omental tumor, retroperitoneal cyst, rupture of an ectopic pregnancy, cholelithiasis, uterine fibroma, and renal tumor had been suspected. Roentgenographic studies, although frequently positive, do not delimit the diagnosis.

The treatment of choice is complete surgical extirpation of the tumor whenever feasible. As there is no definite delimitation between the tumor and the liver parenchyma, complete excision requires sacrificing normal liver tissue. Surgical interference has resulted in cures in 58 of the recorded cases.[11, 12, 13] Following operation x-ray therapy is advised to prevent recurrences.[5] Other therapeutic methods suggested include the injection of sclerosing solutions aiming at the production of thrombosis, fibrosis, and obliteration of the hemangioma.[11]

SUMMARY

1. A diffuse cavernous hemangioma of the liver occurring in a two and one-half year old white male has been reported. There was marked interference with hepatic function resulting in a prolonged bleeding time and tendency to hemorrhage. From our review of the literature, this is the first case of its kind to be recorded.

2. A discussion of hepatic tumors in general with special emphasis on hemangioma has been given.

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CLINICO-PATHOLOGICAL CONFERENCE

Directed by: E. Clarence Rice, M.D.
Assisted by: Francis J. Troendle, M.D.
William M. Crowell, M.D.
By Invitation: Marshall Sanford, M.D.

Francis J. Troendle, M.D.

W. W., a two months old white male was readmitted to the hospital on September fourth because of difficult respirations and cyanosis.

This infant was first admitted at two weeks of age with the same complaint. The birth history showed that delivery followed medical induction two weeks before term for reasons unknown. The infant had almost complete bilateral atelectasis requiring oxygen administration for his first three days of life but responded satisfactorily. He had been perfectly well at home until the evening of admission when there was sudden stoppage of respirations with development of cyanosis apparently unassociated with feeding or vomiting. On physical examination the color was fair, the skin was cold and respirations were weak. His temperature was 97.2 F. Breath sounds were coming through but were considered to be distant on the right side. Roentgen examination showed the mediastinum to be markedly shifted to the right with some compensatory emphysema of the left lung (Figure 1). Reexamination on the third hospital day showed no change by x-ray. On the eighth day roentgen study of the chest showed the atelectatic area of the upper right chest to be aerated and the heart and great blood vessels to be in their normal position. It was noted that a triangular shadow extended from the mediastinum to the periphery of the chest at the fourth interspace on the left side (Figure 2). This was considered by the roentgenologist to be inflammatory in nature. The general condition of the patient improved about the fifth hospital day and he was discharged on the eleventh day.

The child was asymptomatic at home for two weeks when he suddenly became dyspneic and was readmitted. Physical examination showed the child to have inspiratory retraction of the sternum, decreased breath sounds of both lung fields and occasional rales. The admission temperature was 100 F.

Roentgen examination with fluoroscopy revealed a slightly elevated and fixed left diaphragm. There was a uniform opacity over the entire left chest; the interlobar fissure on the right extended across the mid-line to the left, and the mediastinum, in its lower half, herniated into the left chest (Figure 3). These findings were interpreted as indicative of left lower lobe atelec-

tasis. The lateral view showed a mass, extending from the second to the fifth interspace, in the posterior mediastinum, displacing the trachea anteriorly and to the right (Figure 4).

The infant was given a total of 750r to the anterior and posterior chest over a four week period. Following three irradiation treatments the left

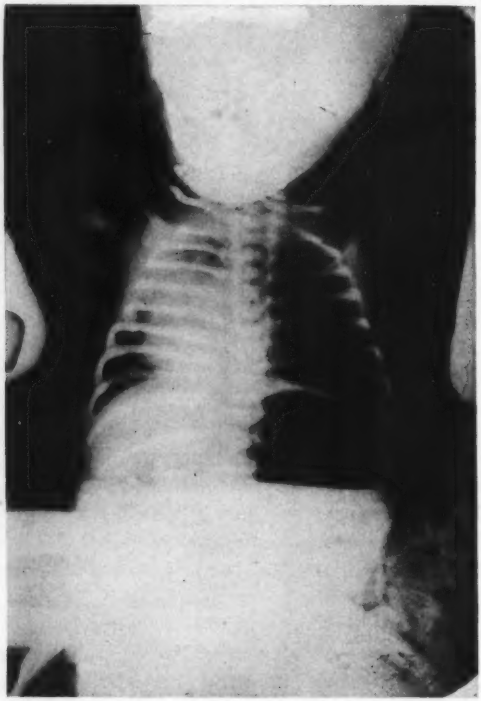

FIG. 1

lower lobe appeared to be reexpanded and the mass in the posterior mediastinum appeared to be somewhat diminished in size and another course of therapy was suggested. Three days later the supra cardiac mass was thought to have undergone further resolution. The patient's course was fair and he was discharged on the 15th day.

Two weeks after discharge roentgen examination of the chest revealed the mediastinal contents to be displaced to the left (Figure 5).

Four days later the infant was readmitted with the history of wheezing respirations since his discharge which was aggravated in the twenty-four

hour period prior to the admission and associated with increasing cyanosis of several hours duration. Just before entry the patient had ceased breathing entirely but was revived with the aid of artificial respiration.

Physical examination revealed a fairly well developed and nourished infant who was restless and had increasing cyanosis while out of oxygen.

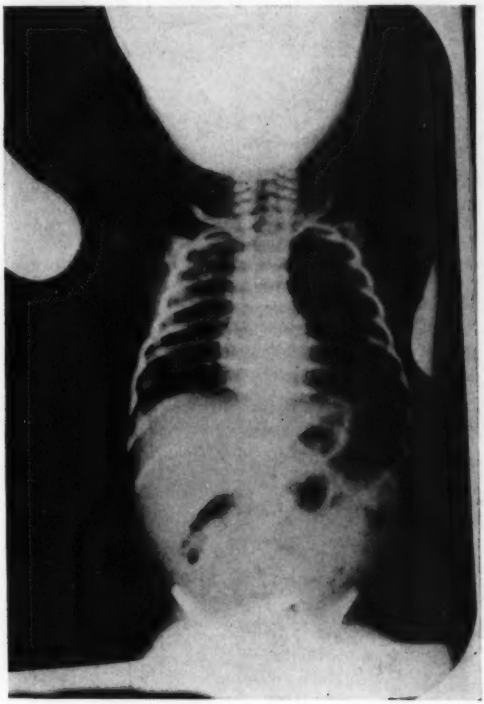

FIG. 2

There was extreme inspiratory sternal and epigastric retraction with dullness over the left lung and absent breath sounds. The heart sounds were regular and of good quality. The abdomen was distended but otherwise negative.

X-ray examination revealed no change from the previous one.

Roentgen examination on the 12th day showed the retropharyngeal space to be normal. The trachea was still displaced anteriorly by a mass which was about the same size as when first noted by x-ray (Figure 6). The heart was slightly displaced to the left.

The infant had increasing periods of cyanosis and difficult respirations. Despite oxygen and stimulants his course was progressively downhill and he expired on the nineteenth day.

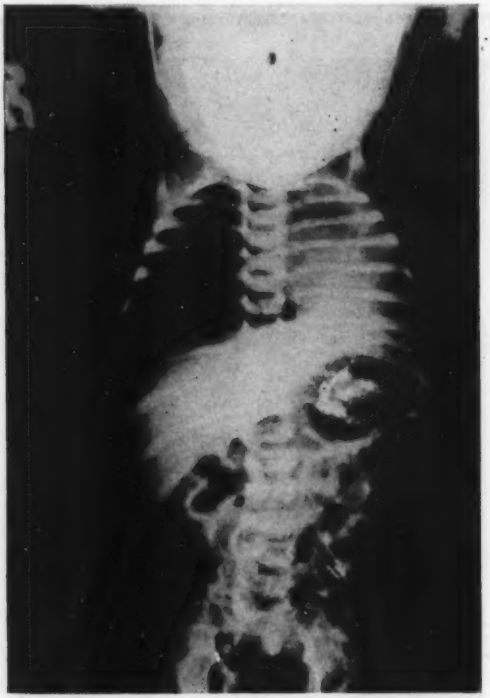

FIG. 3

DISCUSSION

Marshall Sanford, M.D.: To briefly summarize the history, we have a two month old white male infant who was admitted to the hospital because of respiratory distress and cyanosis. These difficulties began at birth, the baby having complete bilateral atelectasis for which he was treated during the first three days of life with oxygen. A satisfactory response was obtained. The patient was then completely asymptomatic for a two day period following which there were recurrent bouts of bilateral intermittent atelectasis first involving the right and later the left lung. These always came on suddenly usually associated with cessation of respirations and unassociated

with feeding or vomiting. A posterior midline, mediastinal mass later appeared; this was treated with x-ray therapy without any appreciable change. Wheezing respirations occurred intermittently. The respiratory distress increased along with cyanosis, and the patient died after a rapidly progressive downhill course.

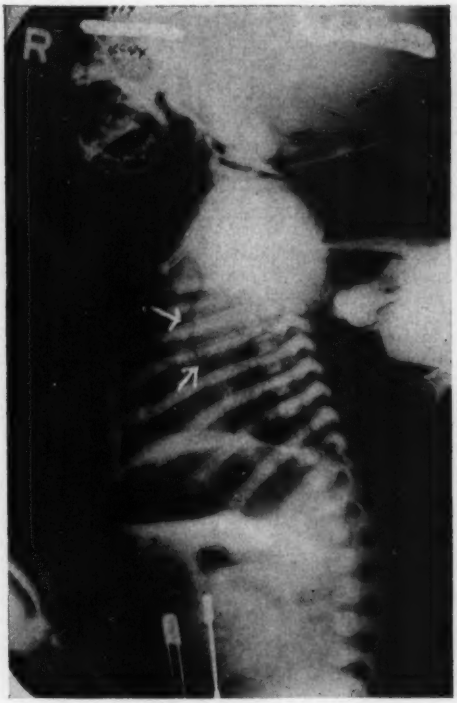

FIG. 4

This is an extremely interesting case and brings up many diagnostic possibilities. The first three days of life could easily be explained by:

1. Persistent atelectasis which is extremely common in premature infants. It usually clears spontaneously in two or three days as in this case. It would explain however, none of the subsequent findings.
2. Congenital malformations of the trachea can be ruled out because of the two months history of difficult respirations. Tracheo-esophageal fistula and tracheal atresia or stenosis cause symptoms early

which usually become progressively worse. They could not explain the presence of a posterior mediastinal mass and these conditions are almost always aggravated by feedings. Papillary tumor of the larynx or trachea could cause some of the symptoms found in this case but not the mediastinal mass.

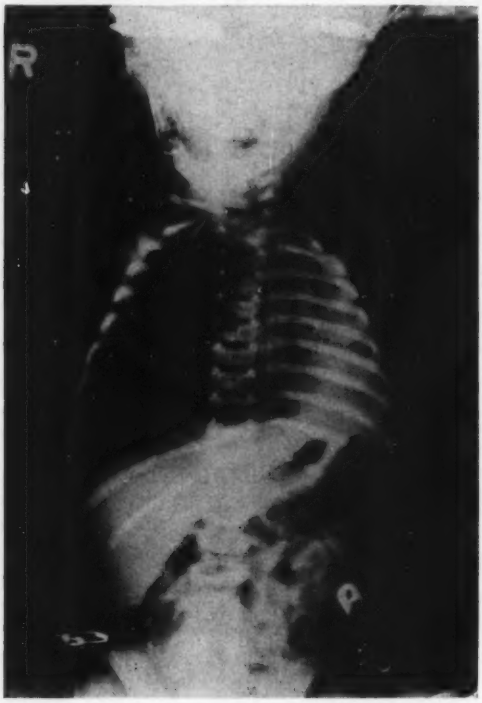

FIG. 5

3. Infection, either pneumonia or bronchiectasis, is considered by many to be the most common cause of bilateral atelectasis, but the suddenness of onset of the respiratory distress and the presence of a mediastinal mass could easily rule out these conditions.
4. Probably because of my interest in the subject, I considered a diaphragmatic hernia as a possible explanation for many of the signs, symptoms, and x-ray findings. However, the persistence of a definite shadow is certainly against this, and since the patient had no symptoms of intestinal obstruction and bowel sounds were never heard in the chest, I think this possibility can be excluded.

5. A lung cyst or cystic disease of the lung would easily explain the x-ray findings but not the subsequent course. As you all must know, most patients with this condition do not have symptoms unless a cyst ruptures, producing a pneumo-thorax and/or empyema. Attention is usually called to this condition by a bout of infection usually thought to be pneumonia, but which is probably

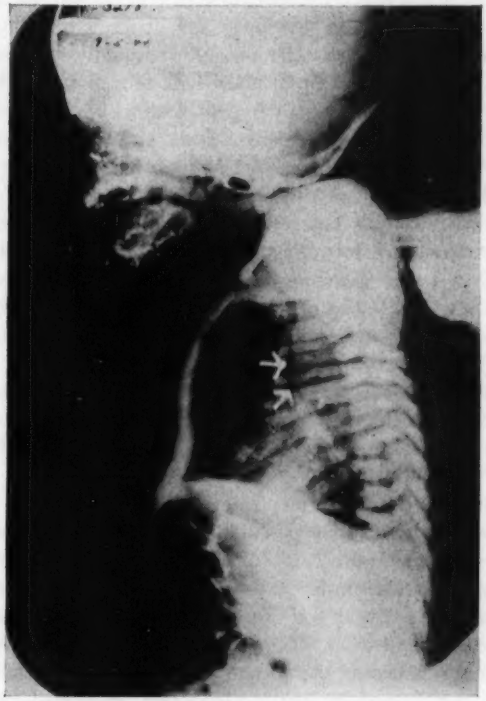

FIG. 6

due to rupture of a cyst. Since this was not the course of this patient and because of the persistent mass, I think this condition can safely be excluded.

6. Benign tumors such as lipomas, fibromas, and chondromas do occur in the mediastinum but they are located in the anterior portion and their growth is so slow that symptoms such as these are rarely produced. Also they are extremely rare in infants of this age.
7. Malignant tumors of the mediastinum are fairly common although this baby is rather young, it has often been said that any solid

tumor in the chest of an infant should be considered malignant until proven otherwise. This type of tumor certainly would be more likely to cause nerve paralysis which this patient subsequently developed. The lymphomas such as Hodgkin's, lymphoblastoma and lymphosarcoma do occur but with great rarity in children so young. Hodgkin's disease is usually associated with enlargement of other nodes and a febrile course is typical. Although lymphosarcoma could occur in this early age, there is usually an anterior mediastinal mass, and the patient frequently becomes malnourished, instead of being well nourished as was the patient we have under consideration. The neurogenic tumors arising from the sympathetic and spinal nerve ganglia are certainly the most common posterior mediastinal tumors, the neurofibrosarcoma probably being the most common at this early age. I have never seen such a tumor cause tracheal compression without first causing esophageal constriction, and since they usually are not encountered in so young a patient I would exclude these tumors from consideration.

8. Dermoids or teratomas are found in patients of this age group but again they are anterior mediastinal tumors that usually do not cause tracheal compression early, and often, though not always, calcification, bone, cartilage, teeth, etc. can be visualized on x-ray. This was not the case in this patient and since they so rarely are found in the posterior mediastinum, I do not believe that they explain the findings today.
9. Duplication of the alimentary tract which is often called by a variety of names is a distinct possibility in this patient since they are congenital and cause symptoms early by gradual increase in size. I recently saw a similar patient with duplication, a mass presenting in the right side of the chest, causing both esophageal and tracheal compression. The cystic structure contained clear fluid, and gastric mucosa. As you undoubtedly realize, these duplications can occur throughout entire alimentary tract and are intimately adherent to the wall of the tract at the site of the occurrence. It is thought that at one time that there was a communication between the lumen of the bowel and the cystic duplication. However, it is almost never demonstrated and since the mucosa may be lined by esophageal, gastric, or intestinal type cells, the secretions vary accordingly. Since the wall of the tube is one and the same, attempt at surgical removal is extremely difficult for no plane of cleavage can be obtained. The treatment of choice is resection of the cystic structure together with the intestinal tract to which it is adherent if this is possible. Since I have never seen a case of

duplication in the thorax which did not first cause esophageal compression, I feel that this condition can be ruled out.

10. Bronchiogenic cyst is a condition which must be considered, and, as I neglected to say earlier, this or any other mass in order to cause the symptoms which we have to explain, would be so located that it would cause tracheal compression in the region of the carina since the atelectasis, at one time, involved the right and later the left lung and depending on the degree of compression, either a partial or complete atelectasis, and/or emphysema would be caused. Bronchiogenic cyst always moves with swallowing. However, no mention of this was made in the history. I cannot remember a bronchiogenic cyst which has caused tracheal compression without associated esophageal compression and for this reason, plus the fact that phrenic nerve paralysis for a bronchiogenic cyst is extremely uncommon, I would tend to exclude this as the diagnosis.
11. The condition which to me best explains all of the findings is tuberculosis of the tracheo-bronchial lymph nodes. This baby is rather young for this to have occurred, however, it is possible. The respiratory difficulty in the first few days of life must be considered, but the persistent atelectasis probably was not associated with tuberculosis. The first bout of sudden respiratory difficulty began at about two weeks of age, which probably was the time that the lymph nodes involved in the primary compression began to enlarge as they do when the allergy first develops. Symptoms come and go but usually occur early and are prolonged. Unilateral phrenic nerve paralysis is common. The dyspnea often is paroxysmal and extremely severe. I am disturbed by the failure of the history to mention a cough since a dry brassy bitonal cough is extremely common in this condition and often is helpful in determining whether or not this is due to caseous nodes causing tracheal irritation or to nerve stimulation. The lymph nodes cause tracheo-bronchial compression since they are closely adherent to the structures and asphyxiation often occurs when the caseous nodes ulcerate into the tracheo-bronchial tree. There it acts as a foreign body causing atelectasis or emphysema depending on the completeness of the block. The location of the involved blockage will account for the changing lung findings. Many of these segments or nodes are coughed up and the patient is asymptomatic again. I believe this explains all the patient's findings and his clinical course.

In summary, I feel that this patient at birth had persistent atelectasis and then developed tuberculosis of the tracheo-bronchial lymph nodes with compression and later ulceration of the nodes into the trachea, resulting

in atelectasis and emphysema. The phrenic nerve paralysis appears to have been caused by direct involvement and compression by the nodes.

Joseph M. LoPresti, M.D.: The changing location of the atelectasis and the periods during which the patient was free from symptoms make me think of one condition, a cyst arising at the level of the carina. This cyst is benign, congenital, and arises from the trachea. Such a cyst could conceivably compress one or other of the main stem bronchi, and thus produce the changing sites of atelectasis. I feel that the correct diagnosis in this case is a congenital bronchiogenic cyst.

Student: What about the possibility of a vascular ring around the trachea?

Joseph M. LoPresti, M.D.: A vascular ring usually produces symptoms of regurgitation and cyanosis during feeding, those of esophageal compression being most prominent at the time of death.

Robert Anderson, M.D.: I suggest the diagnosis of lymphosarcoma.

Joseph M. LoPresti, M.D.: I think that the fact that this child was so well nourished almost rules out the possibility of any malignancy. Would a bronchiogenic cyst in a child of this age be operable?

Marshall Sanford, M.D.: Yes, there is an excellent possibility that a bronchiogenic cyst could be removed easily at operation.

PATHOLOGIC DISCUSSION

E. Clarence Rice, M.D.: At necropsy the body weighed 3.7 kilograms, was well developed, but poorly nourished. The findings of importance were within the thorax.

The thymus was atrophic. A cyst measuring 3.5 by 2.2 centimeters was found in the anterior mediastinum and was located between the trachea and esophagus. It extended more to the left than to the right. The lower end of the trachea and the first one centimeter of each main stem bronchus was attached to the tumor. The left bronchus was compressed by the latter. No communication between the cyst and the air passages could be demonstrated. The right lung was markedly emphysematous, its medial border extending just beyond the left lateral border of the sternum. Slight evidence of edema was found. The left lung was completely collapsed and lay against the posterior thoracic wall. It had a rubbery consistency and was pinkish-blue in color.

The heart lay entirely to the left of the midline. It was normal in all respects. The abdominal viscera revealed nothing remarkable. The cyst contained 8 milliliters of grayish, thick mucus. A rare cholesterol crystal was noted microscopically and no cells were present. The interior was smooth and grayish-white. Histopathologic examination showed the wall to be made up by fibrous tissue with a well differentiated epithelial lining similar to bronchial epithelium.

Anatomic diagnosis was bronchiogenic cyst, with compression of the left main bronchus and atelectasis of the left lung with displacement of the heart to the left.

Difficulty in breathing in a newborn infant is not an unusual finding in the patients of a children's hospital. Congenital anomalies or intrathoracic tumors are the most common causes of these difficulties. Foreign bodies are more frequently seen in older children, after they begin to put various objects into their mouths. Infections may be the cause of dyspnea; however, elevation of the temperature above normal and other evidence of bacterial invasion are ordinarily evident. This child at no time had a temperature over 100.0 F. From the evidence given in the protocol, an acute infection would seem to have been unlikely. Bronchial obstruction due to tuberculous lymph nodes would be unlikely in my opinion in a baby of this age.

Congenital heart disease and vascular anomalies are common causes of cyanosis and dyspnea in patients of the age of the one we have under discussion. Thymic enlargement has been said to frequently cause respiratory embarrassment, however, such a clinical diagnosis has practically never been confirmed by our necropsy findings. We have never seen a malignant thymic tumor at this hospital. The most common malignant tumors of the thorax seen in infants at the Children's Hospital are the lymphoblastomas and neuroblastomas. Of the benign tumors seen in the young child, dermoid cysts, teratomas, lipomas, and congenital cysts have been the most frequently reported, however, none are common. The tumor under consideration is an example of a congenital cyst arising from the respiratory system.

Mediastinal cysts may be of gastrogenic or bronchogenic origin. The former are said by Mixter and Clifford[1] to arise from the primitive foregut before its separation from the primitive trachea. The bronchogenic cysts are thought to originate in the same manner from the tracheal bud. Such tumors may be relatively asymptomatic until they increase in size due to the accumulation of the enclosed fluid and thus compress the adjacent structures.

When the correct diagnosis is made surgical treatment may be effective although the operative risk is great in patients in this age group.

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